Pulmonary Hypertension Association - Ireland

PHéileacán

Newsletter from the Pulmonary Hypertension Association – Ireland: Vol 1 : Issue 2 : January 2019

World PH Day 2018 was celebrated on top of the world by our own PH teddy & friends

Contents:

Message from the Editor

Regina Prenderville
Project Manager, Pulmonary Hypertension Association

Getting Breathless en route to Everest

Welcome to the 2nd edition of PHéileacán. I'm sure you will agree the photos on our cover page are very impressive. They were taken on World PH Day – 5th May 2018. We are indebted to Paddy Brennock who undertook the arduous climb on behalf of the PH Association in memory of his Mother. Paddy was accompanied by a team of 3 other people and of course our PH Teddy Bear. The climb to Everest base camp and to the top of Kala Pather was 18,150 ft. This took 14 days to complete with an average of 8 hours climbing every day. This was a huge achievement by all and a great way to raise awareness of Pulmonary Hypertension.

Tallaght Fun Run

On 6th May members of the Tallaght Athletics Club organised a 5K fun run. This was a real family affair with babies in buggys and pets participating too. We received extensive coverage in the local media for this event.

Ladies Mini Marathon

The Dublin Ladies Mini Marathon was held on 3rd June. As in previous years, we are most grateful to the ladies who choose to support PH on this wonderful occasion. We were delighted to be supported by our old reliables who have never let us down and we welcome the newcomers who have come on board.

Hell & Back

The Hell and Back Challenge was held on 16th June. This is Ireland's toughest physical and mental endurance test. We are most grateful to the 33 volunteers who participated in memory of their dear friend, Addis Doyle. They experienced extreme discomfort while wading through swamps, scaling walls, avalanches of mud and ice baths – not for the faint hearted!!

Cobh Ramblers make history while supporting PH

On Monday, 6th August History was made. Cobh Ramblers (the Rams) Soccer team played a very important semi final football match against Dundalk (who were the current holders of the EA Championship). The Rams were the underdogs and hadn't reached a final in 35 years. However they were happy to promote Pulmonary Hypertension and wore our T shirts on the pitch while warming up for the match. The Rams were victorious with a 1: 0 win. There were enormous celebrations afterwards and great interest was given to our T shirts which were considered 'lucky'

Irish Water Supports PH

To mark PH Awareness week (22-28 October 2018) the staff of Irish Water organised a fun run of 5K through Fairview Park in Dublin. This was held on Tuesday, 23rd October. Seventy five staff members registered to do the run and we are most grateful to them for their contribution. **(See photos of Fundraising Activities on pages 23-28)**

Annual Patient & Family Meeting 2018

Our AGM was held on Saturday 28th April 2018. Many said it was the best meeting we have ever had. This was partly due to the larger audience of 124 but also to the excellent line up of speakers. On the medical side, Prof Jim Egan gave an Update on Transplantation, Prof Kevin Walsh spoke about Intervention in PAH, Prof Sean Gaine and Dr Brian McCullagh spoke about Updates from the world of PH and what we can expect while looking to the future.

Pisana Ferrari (who received a double lung transplant in 2002) gave some very Useful and Practical Every day Tips for those Living with PH, Katrin Hetebrüegge (whose daughter was diagnosed with PH) gave a presentation on a fundraising event named Cycling for a Cure. Patrick Corkery told the story of how he swam the Channel only 20 months following pulmonary endarterectomy surgery. The lectures were followed by 4 workshops. **(See photos on pages 37-38)**

Many have requested that we choose a different time of year for our AGM as the date often clashes with First Holy Communions etc**. We have changed the date for our 2019 meeting to Saturday 7th September 2019 in the Crowne Plaza Hotel, Blanchardstown.**

Patient Support Group Meetings

Meetings are held every 4-6 weeks and we are very pleased to note that many friendships have developed through these regular events. We encourage members to attend when possible. Minutes are produced after every meeting and circulated to those on email.

Forthcoming meetings: Monday 11th February 2019: Clayton Hotel Cork City at 12.30pm
Monday 11th March 2019: Crowne Plaza Hotel, Blanchardstown at 12 noon

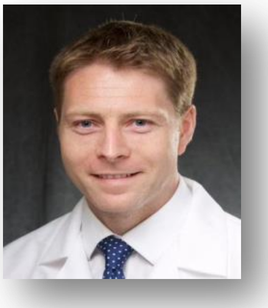
Christmas Lunch: As always our annual Christmas Lunch was a most enjoyable event. This was held in the Crowne Plaza Hotel on 5th December.

Communication: Please note, because of the financial burden of postage, we are unable to communicate on a regular basis through this medium. More than half of our members have submitted their email addresses. If you are not receiving emails from the PH Association – please make contact with us and we will include you on the list.

I hope 2019 will be a happy and healthy year for all of us and I look forward to working with you as we strive to raise awareness of Pulmonary Hypertension.

DEVELOPMENTS IN PH ACTIVITIES IN PULMONARY *HYPERTENSION UNIT, MATER UNIVERSITY HOSPITAL*

Dr Brian McCullagh, Consultant Respiratory Physician



Dear PH Community

The past year has been an eventful one, both from the perspective of the PH unit and the wider community throughout Ireland.

The PH unit welcomes our new clinical nurse specialist Raquel Arean Valle, whom many of you will have met on the wards and clinics. She will be an excellent addition to the team in supporting the service into the future. One of the immediate advantages of having additional clinical nurse support has been the role-out of a joint peripheral clinic with our colleague Dr Des Murphy in Cork University Hospital. This 'Hub and Spoke' initiative has long been an ambition of ours. We will continue this effort into 2019 and if successful other sites such as Limerick or Galway may be considered in the future. This will allow for closer collaboration with our colleagues throughout Ireland, but also makes life easier for those of you that have to undergo lengthy, arduous journeys to visit the clinic in Dublin.

The additional nursing support has also allowed us to partake in 2 multicentre international trials investigating the outcomes of differing drug regimens. We currently have patients enrolled in both trials. The results will take another year to become available but regardless of the outcomes these trials will further our knowledge on the best treatment regimens for PH and we are excited to be involved.

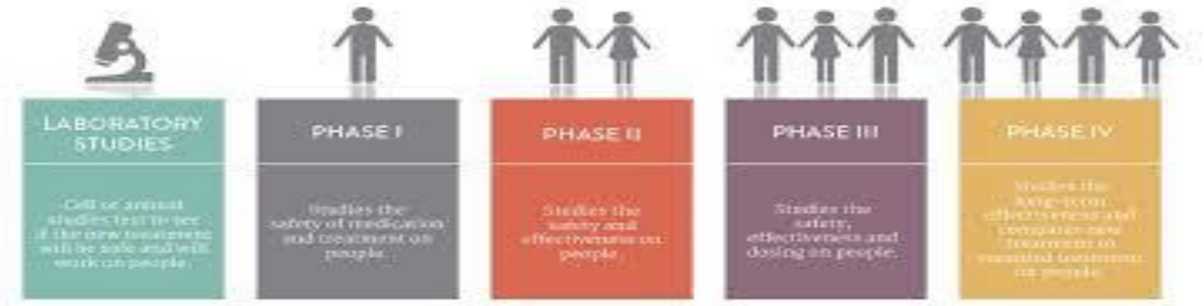
On a local level during 2018 we collaborated with our research colleagues in both UCD and DCU. Many of you will have provided blood samples for our research efforts throughout the years and thank you to the family members who donated samples at our annual PH forum. Your generosity allowed us to complete studies which we presented at an international PH meeting in London in November with positive feedback from our UK colleagues. Furthermore as part of collaboration with DCU we aim to enrol patients very soon into a PH specific home exercise programme (the first of its kind!) the results of which will be of interest to the wider international PH community.

Finally, many of you will be all too aware of the challenges of living with PH, be they social, psychological, physical and/or monetary. While we try to address some of these in the clinic it is impossible to address them all so we remain extremely grateful to the Pulmonary Hypertension Association for the tireless dedication to PH Ireland. The support group meetings are a great source of camaraderie for our patients and it is great to see the level of contributions by patients in this year's newsletter. The annual forum for patients and their families has firmly established itself on our calendar and last year's meeting was of an extremely high standard. We were grateful too, to have the opportunity to make our voices heard in Dáil Éireann in 2018 to highlight many of the above mentioned challenges and let's hope that 2019 brings further positive developments as a result.

Clinical trials in Pulmonary Hypertension

By: Diane Moran & Denise Lennon

Nursing Staff PH Unit



What is a clinical trial?

A clinical trial is a study where the effectiveness of a treatment is tested in a group of people, e.g. people with pulmonary hypertension (PH). Before a medicinal product can be authorised for use, it must go through the clinical trials process to ensure that it is safe and effective and also that the quality of the product is sufficient. Clinical trials begin with small studies in a controlled population of volunteers or patients and, as data is gathered expand to large scale studies in patients. These large scale studies will often investigate the new product and the currently used treatment to see how these two compare. As information is obtained, larger numbers of patients are exposed to the new product and safety data can be collected showing the safety of the product in the intended patient population. Information on the quality of the product and its non-clinical safety will have been obtained before the clinical trial programme commences.

Legislation:

Clinical trials in Ireland are currently governed by the European Communities (Clinical Trials on Medicinal Products for Human Use) Regulations, 2004, SI No 190 of 2004 - http://www.irishstatutebook.ie/eli/2004/si/190/made/en

Why are clinical trials important?

Clinical trials play a very important role in the development of new treatments for different diseases and conditions. Without clinical trials there would have been no developments in new treatments for PH.

Why participate in a clinical trial?

Clinical trials could help you get a new treatment for PH before it is approved. You will also be helping improve medical advances in the treatment of PH. PH is a relatively uncommon disease. This means that it may be difficult for drugs companies to get enough people involved in a clinical trial and can delay new treatments becoming available.

Why to think about before you enrol on a clinical trial?

- Your PH may improve during a clinical trial, but it may stay the same.
- The number of clinic visits may be more than the amount of times you normally attend.
- The drugs that are tested in people with a condition such as PH have already been tested in healthy volunteers. The risk of serious side effects are very low.
- You may be given a placebo (dummy drug) during the trial and not the drug being tested.
- Some of your blood samples may be kept after the clinical trial ends (with your consent).

Clinical trials and PH at the Pulmonary Hypertension Unit:

As the breadth of PH research expands, the Pulmonary Hypertension Unit is committed to participating in ongoing clinical research pertaining to evaluation and treatment of pulmonary hypertension. There are several new drug treatments in early clinical trials, or about to start clinical trials, specifically in people with PH.

An experienced team you can trust:

The Pulmonary Hypertension Unit is well established and has an experienced and committed team working to improve the outcomes for patients with pulmonary hypertension. The Pulmonary Hypertension team includes medical consultants, nurses and administration staff.

Current Trials at the Pulmonary Hypertension Unit

Effect of Selexipag on Daily Life Physical Activity of Patients with Pulmonary Arterial Hypertension. (TRACE)

Brief Summary:

The primary objective of this study is to evaluate the effect of selexipag on the physical activity of patients with pulmonary arterial hypertension (PAH) in their daily life, by using a wearable wrist device (actigraph). The actigraph will collect data on daily life physical activity in the patient's real environment. In addition, the PAH symptoms and their impacts will be assessed by using an electronic patient reported outcome measure in the patient's real environment. Patients will be assigned randomly to either selexipag or placebo.

The Efficacy and Safety of Initial Triple versus Initial Dual Oral Combination Therapy in Patients With Newly Diagnosed Pulmonary Arterial Hypertension (TRITON)

Brief Summary:

The objective of this clinical trial is to compare the efficacy and safety of an initial triple oral treatment regimen (macitentan, tadalafil, selexipag) versus an initial dual oral treatment regimen (macitentan, tadalafil, placebo) in newly diagnosed, treatment-naïve patients with pulmonary arterial hypertension.

Useful links:

1. Health Products Regulatory Authority
2. http://www.hpra.ie/homepage/medicines/regulatory-information/clinical-trials
   clinicaltrials.ie - www.clinicaltrials.ie/

First Hub & Spoke Clinic in Ireland

Caitriona Minnock, Clinical Nurse Specialist

2018 was a landmark year for the National Pulmonary Hypertension Unit (NPHU) with the introduction of the first "Hub and Spoke" clinic in Ireland. A number of combined pulmonary hypertension clinics have been undertaken at Cork University Hospital (CUH) with our colleague Dr Desmond Murphy and the NPHU. This initiative evolved as a result of a number of factors;

- Increasing numbers of complex patients requiring expert input on a more regular basis
- Request from colleagues to provide expert assistance complementing the local approach to care
- Recommendation from the ESC Guidelines for Pulmonary Hypertension

A "Hub and Spoke" approach is not a new model but one which has been adopted by many different disciplines including the business and service industry. This approach involves the expert centre "Hub" providing an array of services that are evidenced as beneficial for the patients complemented by the local centre "spoke" providing regular evaluation and follow up. This model of care seems fitting for a rare disease where expertise can be difficult to access and geographical distance makes attendance only at the hub impractical. The hub and spoke model of care allows for collaboration between clinicians caring for patients locally and the expert centre. It also enables patients to be cared for nearer their homes. Patients will still need to attend the hub for therapies and investigations that may not be available otherwise owing to the rarity of the condition but follow up and further management is then possible locally.

The first formal clinic occurred in June 2018 and a further three have been run since. We were generously welcomed by our colleagues at CUH and were able to get to work alongside them. The experience has been mutually beneficial with both areas learning from the process. Patients were able to access a service for which they may not have been able to avail of in Dublin. Routine follow-ups were then conducted locally thus negating the need for a trip to the capital. We are still learning from our experience in attending CUH and have had all manner of challenges so far (not to mention punctures, traffic delays etc but the hospital canteen more than make a up for these inconveniences !!) We have certainly gained an appreciation of what it takes for our patients to come and see us here in Dublin. More clinics are planned and it is hoped that we will add a further spoke to the hub later this year.

COUNSELLING SERVICE

Mary Burns BSc Psychotherapy and Counselling M11375

Being diagnosed with any medical problem can be a shock. When it's something serious that you're going to have for a long time, it can be even harder to adjust to. It's normal to feel frustrated or upset. Depending on the circumstance of your Pulmonary Hypertension, chances are you'll have to make some adjustments to your lifestyle to manage the symptoms. Whatever you're suffering from, there are some lifestyle changes that you can make that will keep you healthier.

It's vital to learn about your illness. By getting to know and understand what is happening in your body, you'll feel more in control. It might feel difficult to talk about your illness with others. Often they won't understand what it means, why it has happened, or why you can't do everything you used to do. Who you talk to about your illness is totally up to you. Understand that your friends and family have limitations. Sometimes people won't know how to deal with your being sick. Remember that their inability to understand your illness has to do with *their* limitations, not yours.

The support of a counsellor is available to all Pulmonary Hypertension patients and family members. This facility is available without charge, to help you and your family, come to terms with living with Pulmonary Hypertension.

I attend the Pulmonary Hypertension clinic on Tuesday mornings. If this venue or time is unsuitable, it is possible to arrange an appointment in an alternative venue, at a different time. Face time and Skype sessions are also options, for those outside the Dublin area, or for those who are unable to travel.

If you feel you need to talk, or a family member needs support, contact me at the following

085 7631102 email marypburns@eircom.net

Biobanking Update

Clare Hughes PhD, University College Dublin

On behalf of the Pulmonary Hypertension research group in UCD, we would like to extend our deepest gratitude to all patients attending the PH clinic in the Mater hospital who participated in our research study and donated their blood sample.

PH is a condition where the small arteries in the lungs become narrower making blood flow through the vessels more difficult causing an increase in blood pressure in the lungs. Myself and a team of researchers in the Conway Institute, UCD are working on identifying specific proteins which may be responsible for the changes which occur in the vessels. We measured levels of two proteins called CXCL11 and CXCL12 in the blood and found that they were higher in patients compared to controls. Although more work needs to be done, these results suggest that these proteins may potentially serve as biomarkers in PH. A biomarker is a biological protein which may indicate the presence of a disease, may provide clues as to the mechanism of the disease or may demonstrate how well patients are responding to treatment. The results of this work were recently presented at the pulmonary hypertension physicians' research forum in London.

Each of you were one of fifty-three patients from the PH clinic who participated in the study, and in doing so, allowed us to perform important research which may help us to better understand this disease. In addition, we would like to sincerely thank the family and friends of patients living with PH, who attended the Annual meeting on the 28th of April of this year at the Crowne Plaza, and donated their blood sample. These samples were used as control samples for our study. We were taken aback by the amount of people willing to volunteer that day and we are extremely grateful for their contribution.

We really value the support of everyone who participated, and it is only with the help of volunteers like yourselves that research studies like this one are possible. You have made a contribution not only just to our research efforts but to the field of PH research. Our understanding of the disease has improved greatly over the past 25 years, however we still have a lot to learn. Every piece of information researchers obtain may help to shed light on pulmonary hypertension, its progression and how to treat it.

FLU VACCINE

Patricia Ging (BPharm, MPSI, M.Sc, FFRPS MRPharmS)
Transplant / PH Pharmacist
Pharmacy Dept. Mater Misericordiae University Hospital

Every year in Ireland over 100 people die from flu.
We strongly recommend all of our patients with PH to get the flu vaccine. It is the best protection against the flu which can also cause very serious illness in people with heart or lung conditions. That is why the vaccine is especially important for people with PH. Ideally you should get the vaccine as soon as it becomes available in September but if you have not got it yet then there is still time to protect yourself. Make an appointment with your GP or pharmacy today!

I got the vaccine last year- why do I need it again?

The viruses changes each year. This is why you need to get a new vaccine each year.
The seasonal flu vaccine protects against the 3 strains of flu virus recommended by the World Health Organization (WHO) as the strains most likely to be circulating this season The flu vaccine is free if you are in an at risk group but you may be charged a consultation fee, unless you have a medical card or a GP visit card.

My friend said that she caught the flu from the vaccine- is this true?

No - the flu vaccine doesn't contain any live viruses – therefore it cannot possibly give you the flu. The vaccine does take about two weeks to work- so you are still at risk of catching flu for a while after being vaccinated. You can also catch other viruses.
The flu vaccine helps your immune system to produce antibodies to the flu. If you have been vaccinated and you come into contact with the virus, these antibodies will attack it and stop you from getting sick.

Is there anything else that I can do to prevent infection?

Consider if your spouse or partner or carer should also get the vaccine.

The vaccine is strongly recommended for people who:

- are 65 years of age and over
- are pregnant
- have a long-term health condition e.g. heart, lung, kidney or liver disease or diabetes

- work in healthcare
- are a carer
- live in a nursing home or other long-term care facility
- in regular contact with pigs, poultry or water fowl

Remember If you do get a viral infection with coughing or sneezing "**catch it, bin it, kill it"** this will help to slow the spread of virus in the community

Germs spread easily. Always carry tissues and use them to catch your cough or sneeze.

Germs can live for several hours on tissues. Dispose of your tissue as soon as possible.

Hands can transfer germs to every surface you touch. Clean your hands as soon as you can.

Are there any reasons not to get the vaccine?

Don't get the flu vaccine if you have had a severe allergic (anaphylaxis) reaction to a previous dose or any part of the vaccine.

Vaccination should be re-scheduled if you have an acute illness with a temperature greater than 38°C.

For more information check the website https://www.hse.ie/eng/health/immunisation/pubinfo/flu-vaccination/about-the-vaccine/

Information is also available from your GP or local pharmacy

What can I do if I get viral illness?

Because of your underlying condition and medication you need to be careful not to take anything that could worsen your PH or make your medications less effective. Simple remedies such as paracetamol for headache and shivers are best. If your nose is blocked try a sinus rinse or antihistamine such as loratidine or cetirizine. **Do not** take decongestants as these can cause heart and blood pressure problems and can be dangerous for people with PH. **Do not** take herbal remedies as there could be interactions with your other medicines.

EXERCISE HOME BASED REHABILITION PROGRAMME AS AN ADD-ON THERAPY FOR PULMONARY HYPERTENSION

Ciara McCormack, PhD Researcher, Clinical Exercise Physiology, DCU

Exercise & PH

It’s widely known that exercise, at any stage in life is very beneficial for maintaining cardiovascular health and muscular fitness, improving mood, controlling weight and lowering the risk of high blood, heart disease, dementia and other neurodegenerative diseases. Substantial evidence now exits to support the benefits of exercise training and physical activity (PA) as a treatment option for many chronic diseases such as diabetes, high blood pressure and coronary heart disease.

Patients with PH have difficulty with exertion making it a challenge for patients to know the right type and quantity of activity to undertake thus leading to the avoidance of exercise. Recent research findings indicate that carefully prescribed exercise can be safely used in PH patients and can significantly enhance quality of life and increase functional capacity, allowing activities of daily life to be performed with more vigour and vitality. The first clinical study to examine the safety and feasibility of individually adjusted, supervised exercise in PH patients was published in 2006. A number of subsequent studies have found that exercise training can improve exercise capacity, quality of life and muscle function in PH patients. Recently published guidelines recommend supervised and closely monitored exercise and lung training as add-on to standard medical therapy for stable PH patients. Because the severity of PH differs from patient to patient recommendations on exercise are also different for each individual, and may change over time depending on symptoms and response to treatment.

The researcher & The Programme

My name is Ciara Mc Cormack. I am a Clinical Exercise Physiologist and PhD researcher who specialises in chronic illness rehabilitation. In 2015 I graduated with a first class Honours Degree in Sports Science and Health from Dublin City University (DCU). During my undergraduate studies, I developed a keen interest in the use exercise as a treatment to improve health and quality of life in people with one or more chronic diseases.

Following the completion of my undergraduate degree, I was offered and accepted a Master’s degree scholarship to work on the a project titled PATHway (Physical Activity towards Health), under the supervision of Professor Niall Moyna, an expert in Exercise

Cardiology. PATHway is a technology enabled programme that is designed to promote home-based exercise in people with cardiovascular disease. More recently, I was offered a PhD scholarship that will involve the design delivery and evaluation of a home-based exercise and lifestyle program for individuals in PH. My supervisors are Professor Moyna, Professor Sean Gaine, and Dr. Brian McCullagh Consultant Respiratory Physicians, in Mater Misericordiae University Hospital, Dublin. This is a very exciting project and one in which I am looking forward to starting in the near future.

Last November, I had the privilege of travelling to the beautiful city of Heidelberg in Germany to work with some of the leading international experts in exercise and PH and to see their very successful in-patient PH exercise programme in action. During the visit, I had an opportunity to meet with Professor Ekkehard Grünig, Head of Centre for Pulmonary Hypertension at Heidelberg University Hospital. We discussed the development of a modified home-based exercise programme for Irish PH patients, using their programme as a guideline. With guidance from Professor Grünig and input from Professor Gaine and the team in DCU, we have designed a study to examine the benefits of a home-based exercise programme for PH patients in Ireland.

The study will be 28 weeks long, and will consist of 4 phases. Blood samples, physical fitness, lower and upper body strength, body weight, physical activity levels and quality of life will be measured throughout the study. Phase 1 of the study will be 12 weeks in duration. During this phase, the study participants will continue as normal with no input from the research team. Phase 2, the induction phase will take place during weeks 13 to 15. PH patients will visit DCU/ Mater hospital one day per week during weeks 13, 14 and 15. This phase will focus on exercise and respiratory training, strength work, core/flexibility exercises along with lifestyle/behaviour change. Emphasis will be placed on education regarding safety considerations, self-monitoring of exercise intensity and awareness of relevant warning signs and symptoms. Participants will also be provided with reading and video material to examine, and will be assigned a progressive series of tasks to undertake between scheduled classes. The intervention phase (Phase 3) will commence on week 16 and will involve participants undertaking an individually designed, 12-week home-based exercise program. The number of steps taken per day along with heart rate and a daily diary will be used to monitor daily physical activity. Participants will also receive a weekly phone call to track progress and to advise on exercise goals and prescription. During week 28, participants will return to DCU/Mater hospital for the final assessments.

We are very excited to start this project and we aim to enrol patients very soon. The study is unique and in addition to Irish patients, the results will also be of great interest to the wider international PH community.

Top Left: PH Patients during their exercise session. Top Right: Thoraxklinik Hospital , Heidelberg. Bottom: Ciara McCormack & Prof. Ekkehard Grünig, M.D, Head of Centre for Pulmonary Hypertension at Heidelberg University Hospital, German

Attendance at International Pulmonary Hypertension Conference & Scientific Sessions, Orlando, USA

June 28 – July 1, 2018

PHA Pulmonary Hypertension Association
Empowered by hope

Our mission is to extend and improve the lives of those affected by Pulmonary Hypertension.

Regina Prenderville, Pulmonary Hypertension Association - Ireland

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It was an honour to be invited to attend the above conference which I found informative, inspiring & encouraging for the future. The theme of the meeting was **'PHinding your hope'**

There were many breakout sessions and workshops to attend. The following are some of the notes I took while attending these sessions – (all of my notes are available on our website – www.pulmonaryhypertension.ie)

1. Tips & tricks for conserving your energy:

This session was conducted by two adults with PH and are both support group leaders. The opening comments were 'Life goes on after diagnosis'. PH patients and caregivers need to get the most out of each day by conserving energy, making "me time" and simplifying daily tasks'

Objectives:

- Learn how to cope with constant demands on your mind and body while living with a chronic disease
- Gain an understanding of the ways you exert emotional, mental & physical energy during everyday activities
- Learn how to make life easier

Key Messages:

A person with PH who runs a busy family home only needs 3 things ……. a Maid, a Chef and a Chauffeur!!

There are various levels of activity:

Level 1-2	Brushing your teeth – general conversation
Level 2-3	Getting dressed – cooking
Level 3-4	Showering, washing dishes
Level 4-5	Changing the bed linen

It is estimated that the energy used while showering or washing dishes is the equivalent to walking 3 miles per hour and changing the bed linen is the equivalent to walking at 4 miles per hour

- The best was to conserve energy is learning to say “no”
- Incorporate rest into your schedule – plan for rest, occasionally give yourself a “day off”
- Use oxygen and mobility devices as prescribed
- Sit when possible while doing daily chores or in the shower
- Get comfortable with computers by shopping online, paying bills on line and keeping in touch with friends through social media
- Avoid bending over at the waist towards the floor; use long-handled devices such as a reacher/grabber and sock aids.
- When taking a shower – keep the room ventilated and don’t let steam build up. The shower should be warm, not hot or overly long.
- Alternate motions that use the arms. Avoid holding your arms overhead for long periods of time.
- Push heavy items along a counter versus lifting and carrying
- Keep frequently used items between your waist and shoulder level
- Plan ahead to avoid rushing (eg if going somewhere early in the morning – shower the night before)
- Don’t be afraid to ask for help, use handicap parking facilities
- Know your limitations but still try to maintain your independence
- Finally learn to accept your limitations – this will conserve your emotional energy

2. Communicating in a PH Emergency: Am I prepared?

This session was facilitated by an adult with PH who is a support group leader and told of her medical emergency experience – when she wasn’t prepared.

Objectives:

- Define PH emergency & understand why immediate action is critical
- Learn how to be prepared & proactive in a PH emergency
- Learn how to respond to a PH emergency with confidence as your own advocate

Medical emergency experienced by speaker: This session was facilitated by Dawn Jones who related her experience to the audience. One hot summer’s day Dawn was at the beach with her daughter and grandchildren. She decided to paddle in the water and had her pump in her hand. Unfortunately she twisted her ankle on a stone and fell. The pump became submerged in water and although she immediately got out of the water and tried to dry the pump – the alarm started to ring and the pump failed. Her daughter rushed her to the nearby hospital. Dawn tried to explain PH to the doctor on call. She was hysterical trying to explain how important it was that her IV PH medication cannot be stopped – the doctor tried to sedate her and took her phone from her (she was trying to call her family doctor). There was a very long story about security removing her daughter from the hospital etc & Dawn jumping off the trolley and being man handled by security when ………….thankfully a nurse in the Emergency Room heard the commotion and asked what was going on – she had experience of dealing with PH before and advised the team on the appropriate procedures. Dawn received IV treatment and thankfully made a full recovery.

Preparing for a PH Emergency:

- In the past patients were told to always have their documents with them, now everything can be stored on a mobile phone
- Save your doctor's contact details on your phone
- Scan your latest prescription and relevant documents
- For continuous infusion therapy, know who to contact for pump-related issues & have emergency contact details listed
- When leaving home – bring a spare pump and extra medication with you
- Should your phone be locked - allow the facility to call ICE (in case of emergency). Ensure the person you propose knows your condition and can unlock your phone
- Educate your family, friends and others on how to respond if you have a PH emergency. You could have emergency drills for your home, like a fire drill.
- It is vitally important that whoever your emergency contact is should be fully aware of PH including list of medications and your PH care team's contact information.

Conclusion: The above story was a very serious incident and Dawn was traumatised from the experience. However when she related the situation to her PH nurse, they decided between them that they would not write letters of criticism to the hospital and media etc but would be pro-active. The PH nurse wrote to the Director of Nursing in the Hospital and praised the intervention of the nurse who came to Dawn's rescue. She also wrote to the Medical Director advising him of the incident and proposed that she would arrange an education session for the staff in the ER. She obtained sponsorship and organised an intensive educational session for the staff to ensure a situation like this will never happen again.

3. Living a new normal

Session description:

When diagnosed with PH, you experience so many changes that you may feel like you're no longer living your normal life. Three adult PH patients shared strategies on how to cope with the sadness that comes with losing aspects of your old life as well as adjusting to living a new life and accepting the new you…..

Objectives:

- Understand how even if PH brings on a new normal, you are in control of defining what that normal can look like
- Understand that a new normal is possible and can be wonderful
- Learn how to find creative and safe ways to do what you enjoy

Key Messages

- It's OK if PH rocks your world at first, but the truth is that this disease does not define YOU
- Creating a new normal means figuring out what you can and cannot do anymore – how you can still do things you love, even with PH limitations
- You have a choice whether you have a healthy or unhealthy new normal: ***A healthy new normal*** requires you to focus on the things you can control – such as your diet, exercise, compliance with doctor's orders, self-advocacy with medical teams, self-education about PH – so you feel empowered. ***An unhealthy new normal*** fixates on things outside your control and encourages feelings of helplessness, sadness, frustration and anger.
- Be gentle & patient with yourself, finding a new normal may take time and everyone's journey is unique
- You are not alone when experiencing the frustration of the new normal; reach out to others through support groups
- Remember it's ok to have bad moments – but not ok to fixate on those feelings

Closing Ceremony

Keynote speaker: Rebekah Calverley, young adult with PH

Rebekah shared her journey from being diagnosed with PH as a toddler and how she found ways to cope with the limitations she was presented with. She spoke of how determined she was to live a full and rewarding life. She became an opera singer, an equestrian, an international corporate banker and a PHA fundraiser.

Rebekah told the story of how she was born in Auckland, New Zealand. She was unwell a lot as a baby and when she was two and a half she was diagnosed with a large hole in her heart and her parents were told it was unlikely she would make it to 5 years of age.

When she was eight, her family moved from New Zealand to New Jersey with her Dad's job transfer. She became a patient of Dr Diane Kerstein at the Paediatric PH Centre at Columbia University Medical Centre, which was one of the nation's first PHA accredited centres of Comprehensive Care.

Prior to moving to the US, Rebekah was a keen equestrian, loved singing and spending her days on the ocean. When she was a young girl she was conscious of her limitations but continued trying to keep up with her class mates in the various school athletic activities. She used to run sprints during PE or try to walk and talk – however she would always end up in the school nurse's office on oxygen etc. She eventually accepted her limitations. Within the constraints of her illness, she continued horseback riding for several years and began taking voice lessons, performing opera recitals and singing in her church choir.

In college, she majored in Finance and Accounting with a minor in Marketing. She was Student President of the Business school and worked several part-time jobs. She has recently moved into a middle management position with an Australian bank in New York.

Rebekah spoke very emotionally about the support given to her by her family. Her parents Jacinta & Richard have always been there for her and supported her in everything she wanted to do. Because of her determination and stubbornness – her Mum often said ‘God made your head stronger than your heart’. She described her sisters as her ‘Bodyguards, her Entourage and her Protectors’. Nothing was impossible when they were growing up – if Rebekah was unable to keep up with others, her sisters just gave her a ‘piggy back’ – To this day they are there for her, looking out for her at all times.

She ended her wonderful & emotional presentation with an amazing recital of Puccini’s ***‘O Mio Babbino Caro’***

Old friends meet at conference

One of the many exhibition stands where staff were most helpful and friendly

Michael Knappen,Director Patient & Caregiver Programs , PHA, celebrates with delegates at the opening of Fashion Show at 2018 Meeting

Pulmonary Hypertension Association - Ireland

Patient Activities support PH Association

We greatly appreciate the support given to us by our patients, their families and friends who raise funds for our Association. Through your generosity we are able to support the following activities:

- Annual conference for patients and their families
- Regular pulmonary hypertension awareness events
- Regular support group meetings giving patients the opportunity to meet and learn
- Grants for trainee doctors & nurses to increase their expertise in PH
- Guest speakers at support group meetings
- A counselling service for PH patients
- Newsletter
- Website, containing information, news.
- PH Merchandise

The following pages give an insight to the commitment and dedication of our many supporters. Should you decide to organise an activity on our behalf – do remember to take some photos and send them to us for inclusion on our website and in our newsletter.

World PH Day 2018

Where to in 2019 ?

Paddy Brennock celebrates World PH Day on 5th May 2018

LADIES MINI MARATHON 2018

Cobh Ramblers take on the Champions and Win !!!

Christine Coakley from the Pulmonary Hypertension Association with members of the Cobh Ramblers team prior to their great victory over Dundalk. Photos courtesy of Sean Truscott Photography

Irish Water Supports PHA - Ireland

Hell and Back Challenge

In memory of Addis Doyle

Tallaght Fun Run

LIVING WITH PH - PATIENT CONTRIBUTIONS

Mindfulness

By: Karen Doherty

I have always practised mindfulness meditation, but recently decided to undertake a nine week beginner’s course, as a refresher.

For those of us living with PH, it can be challenging to stay positive. Physically and emotionally, stress is a big part of our lives. I find it very important to disconnect from my difficulties and just try to connect with myself.

During the course, as each week went by, we learned longer meditations, walking meditations and self-awareness. Speaking within the group at the beginning of each class, about how our meditations helped us the previous week, was very helpful. Listening to other peoples experiences was very interesting and enlightening. Mindfulness helped everybody in the group in a different way, as we were all there for different reasons.

If you are unable to take on a course, there are plenty of guided meditations on YouTube. To help reduce stress and learn to relax, I always found listening to somebody guiding me through a meditation very helpful - especially at the beginning.

Choosing somewhere in your home that's comfortable and quiet is a good idea. Candles can also contribute to a nice 'mood'. I know scented candles are not good for us, so maybe battery operated ones would be a better choice. Short guided meditations are better when beginning. About ten minutes. Just ease yourself into it at first.

Practising mindfulness is a good habit and I would highly recommend it to anybody. It becomes part of your daily life after a while. Why not give it a go!!

Wishing you all my very best wishes for the New Year.

Karen.

A PATIENT'S STORY

By: Michael Sammon

In Autumn 2014 I noticed that when I was out my regular cycles, I was becoming very breathless. Occasionally I would go walking in the Wicklow mountains & again the shortness of breath limited me severely. In a very short space of time, I found that from being very fit to find I was unable to do the simplest of tasks. I decided to have the old body checked out. Our GP - Dr. Adrian McGoldrick had me hospitalised in 2015

After 8 days I was discharged & unfortunately I was no better. I was back in hospital 5 months later. In between times I tried cycling or running as I would not accept I was in bad shape & I know I pushed the *old ticker* to past the limit more than once !!

Time moved on and I was going downhill rapidly - but I was still battling. I got an appointment in the Beacon & met Dr. Robert Kelly. He confirmed that given my age (72 at the time) that my heart was good. This was confirmed by a cardiogram which was carried out in Tallaght Hospital - so I was told I had a respiratory problem. He arranged for an MRI in the Hermitage Clinic & said he would contact a specialist in the Mater Hospital.

Time passed as we waited for an appointment. We were so delighted to receive a call from Prof Sean Gaine's secretary informing us that she could give me a cancellation appointment. I/we had many visits to 56 Eccles St. where the PH team started the interim treatment for the disease, they put me on a course of Adempas/Riociquat tablets which I found a great help.

Down the road Papworth Hospital was looming, I had the necessary tests in the Mater requested by Papworth (UK) and subsequently we went there. We had an interview with the surgeon Mr David Jenkins who told us we just had to wait. However Prof Gaine felt the wait was too long and during one of my visits he contacted Papworth to outline the severity of my condition and I was eventually seen on 28th February. I travelled to the UK with my wife Ann and daughter Elaine. Mr John Dunning performed my surgery which was an outstanding success.

On my return home I had to stick with a strict recovery schedule. So right now I'm doing very well, thankfully to all in Papworth & Prof. Sean Gaine and the PH team in 56 Eccles St. Dublin. I am able to walk and cycle (not too far). I (religiously) attend Ms.Karen Cradock's Cardiac Physio clinic every week.

I am one of the fortunate PH sufferers & I know what you are all going through and I dedicate this article to you. I will continue to keep you in my prayers and I enjoyed meeting you all at our Christmas lunch in December. I know all of our PH people are extremely grateful to those who work behind the scenes for our wellbeing. We also acknowledge the PH Association. Attending the regular meetings is a great way to support each other and keep in touch.

MY EXPERIENCE WITH PH

Anonymous

While we have had many shades of Government over the years, generally all of them have put extra money into our Health Services. The idea is to support the sick and those who care for them. I personally do not wish to make any complaint against our health service or the people who deliver that service, as I have received excellent care from the doctors, nurses, and backup staff whether they be radiographers, porters, kitchen staff, and also the home help services provided by public health nurses, physiotherapists, and dieticians.

I eventually found it necessary to retire from full time employment [on medical advice], about ten years ago following a lengthy hospital stay. Having been diagnosed with SLE twenty four years ago I was able to continue working for fourteen years but it eventually began to affect my organs. A cure has not yet been discovered so right now all that can be done for me is to control its symptoms.

I was granted a medical card on the grounds of having an incurable disease. About the same time I was referred to Prof Sean Gaine in The Mater Hospital. I had been diagnosed with a heart and lung problem. Prof Gaine had a number of tests carried out, which I expect you are all familiar with, the result of which led to me being diagnosed with the now familiar PH. At that time he felt that it was at a reasonably early stage. While the latest medicines for controlling the condition have not been successful for me they are a great help to many patients.

My only treatment now is oxygen, which I also take at night which is provided by way of a concentrator. I feel that the HSE should support the cost of running this machine which is quite expensive. Maybe the powers that be will soon take notice and give us a helping hand.

I would like to thank all the good people who take care of us, Prof Sean Gaine, Dr Brian McCullough, the nursing staff in the PH Unit and all of the para medics who keep us going. Suffering in silence can eat away at you but coming to our occasional meetings organised by the Pulmonary Hypertension Association can be so rewarding. You know that a problem shared is a problem halved. Together we can support each other and we look forward to better days ahead.

Ramblings By M.A.G

IRISH REPRESENTATIVE ON EUROPEAN RESPIRATORY SOCIETY TASK FORCE

Patrick Corkery, who presented at our Annual patient conference in 2018 on his PH experiences and recovery from pulmonary endarterectomy surgery, was subsequently asked to tell his story to a number of European medics in a webinar for the European Respiratory Society (ERS) alongside Prof. Marion Delcroix and Dr. Gergely Meszaros.

As a positive patient role model with his story covering PH illness, treatment, surgery and ultimately culminating in swimming the English Channel, he has also accepted an invitation to be a Patient Representative on the ERS Task Force. Run under the direction of Prof Delcroix, its aim is to publish a Statement Paper on chronic thromboembolic pulmonary hypertension (CTEPH) within a maximum of 2 years. As the only Irish representative and non-medic on the taskforce, he hopes that by sharing his experiences it will assist the medical practitioners in understanding patients' views.

Patrick is already planning his next big swim, in Ireland, the logistics of which are still being finalised. The actual date will be for a summer swim, after the sea has warmed up again. At a similar distance (33km) to the English Channel it is likely to take anywhere from 12-15 hours, depending on tides and weather conditions on the day. He hopes to have a tracker on the boat which will plot his course in real-time and can be followed online. More details to follow.

To give an idea of Patrick's past swim experience, we are reproducing here an article written by Orla O'Muiri and reproduced with kind permission from Outsider Magazine.

What did you dream of doing when you were a 7-year-old kid? Take a minute. Have a think. Maybe it was something like representing your country in the Olympics, or perhaps it was something less glamorous like working in finance?

Patrick Corkery dreamed of swimming the **English Channel**. He did everything to make sure one day that dream would become a reality. Over the years, he slowly and steadily built up his swimming resume so that now it reads rather impressively. It is filled with four decades of swimming and lifeguarding on three continents. It includes marathon swims like Cork to Cobh, Lough Sheelin, Cork Distance Week, Lake Zurich and the Manhattan Island Marathon Swim. Some standout details on it include a top spot on the podium at the Dun

Laoghaire Harbour Race, having two Ice Miles under his belt and two decades of masters galas and open water swims in the Leinster Open Sea races on it.

The final hurdle before realizing that childhood dream was a beast of a different nature, The North Channel, the stretch of water between Northern Ireland and Scotland. In September 2014, a spot opened up and Patrick jumped on it, flying home from his family holiday in Spain for 48 hours to make a stab at it. That's when things started to take a turn for the worse. The North Channel Lion's Mane jellyfish are notorious, ranging in size from a dinner plate to a dustbin lid, and they came out in force to greet Patrick.

"I started getting stung after the first hour. They were like nettle stings, as in they were manageable enough individually but you were getting stung constantly. Normally you get a sting and get out. The first one of the day was on the nose. Later in the day, I got the whole rest of my face where the beard would be but not the nose. And then they'd get one arm, the other arm, the leg, the chest, the other leg, etc.," says Patrick.

"At the end of the day, it boils down to what you want more. Is the pain you are suffering enough to make you want to get out or make you want to finish it more? I was thinking, I'm freezing but I'll be on a sunbed in less than 24 hours and if I get out now I'll be berating myself for not having stuck it out for a few hours longer. I'll be giving out about 31-degree heat in Spain so what's wrong with the 14-degree water I'm in now?"

Patrick managed to talk himself into keeping going, one stroke at a time for ten solid hours. It was then that things started to take a turn for the worse. "I started coughing. I was struggling with breath but part of it was the cold. I was becoming conscious of it the more I swam on. My arms were struggling to turn over. It's the closest I ever felt to drowning.

> "I saw Donal get into his togs. He swam over to me and he touched me and said, 'Your race is over, I've touched you. You have to get out."

"One of the crew, Donal told me to do 200 strokes on my front and 10 on my back. It was something to aim for, so I did it. The second rotation though I could only manage ten on my front and ten on my back. I just couldn't keep my head in the water. I couldn't turn my arms and the boat was drifting away. I said to the crew, 'I need you to help me'. I wanted a support swimmer in with me but I couldn't explain it. I saw Donal get into his togs. He swam over to me and he touched me and said, 'Your race is over, I've touched you. You have to get out."

“My first reaction was, ‘What the f**k? I’m a mile away.’ I really thought I was going to make it. But I knew I was struggling. He had done it this way so as to take the decision out of my hands. I have them there to look after me, so I climbed out. I was shattered about what they had done but I was also in agony. It’s like been sent off in an All-Ireland final. The referee makes a decision and it’s out of your hands. You just have to go.”

Feeds in the North Channel

And just like that, Patrick's North Channel Swim was over. However, the ramifications of attempting it were unfortunately just beginning.

"I went to the doctors that day. I couldn't sleep and I was getting sick. It turned out that I had fluid on my lungs. Three months later, I was told that I had blood clots in my lungs the size of satsumas. For six months after that, I couldn't even lift my little fella up. He was three at the time. The right side of my heart was twice the size of the left side because it was under pressure trying to pump past the blockages.

"In January 2016, I went for double open lung surgery (pulmonary endarterectomy). It means cutting open the sternum and scraping out the residue of the blood clots that hadn't dissolved. It was a 10-hour operation and meant two weeks in the hospital. I went from swimming 20 miles in a day to not being able to walk down the corridor and not lifting anything heavier than a coke bottle. I could be on blood thinners for the rest of my life."

A single decision to take up a cancellation spot to swim the North Channel as a training exercise on the path to his ultimate goal of doing the English Channel instead turned into something that irrevocably altered his life. Yet, strangely, he doesn't regret the fateful decision. "It didn't work out but it was the opportunity of a lifetime."

And the dream of swimming the English Channel remained cemented firmly in Patrick's mind. "My goal was the English Channel. I equate the English Channel to Everest and the North Channel to K2. K2 is seen as harder, technically a much more difficult climb but the one with all the cachet is Everest. If I didn't get to do the English Channel,that would have upset me."

So, on the afternoon of 26 September 2017, 20 months after having open lung surgery, Patrick Corkery finally did it. He swam the English Channel in a time of 13hrs 12mins. Only two out of eight solo swimmers attempting the crossing that day were successful.
"I reached land. I got out and kissed the ground and blessed myself. A lifetime goal had been achieved and it no longer hangs over me."

The English Channel

(Written by Orla O'Muiri of Outsider Magazine and originally published on www.outsider.ie)

AGM 2018

Pulmonary Hypertension Association - Ireland

Join our PH Association today

No registration fee – just forward your name and email address to pha@mater.ie or complete form below and mail it to us

Benefits of being a member:

- Information regarding support services available
- Notification of Support Group Meetings
- Attendance at meetings which patients find very helpful and sociable
- Regular update emails
- Details of activities in PH UK, PH Europe and PHA USA
- Fundraising ideas and start up packs

Our Association will develop and grow through increased membership

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I would like to join the PH Association:

Pulmonary Hypertension Association - Ireland

Name: ____

Address: ____

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Email: _____________________________________ **Telephone :** ___________________

Are you a patient: ☐ **Carer:** ☐ **Parent:** ☐ **Medical Professional:** ☐

Please complete form and return it to PH Association – Ireland (see address below)

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